LEFT HEART HYPOPLASIA WITH ASSOCIATED ANOMALIES

BY

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The malformed heart to be described was removed at necropsy from a 9-day old Bantu infant, and sent to the Department of Anatomy for study. Unfortunately neither the clinical nor necropsy records could subsequently be traced and the only information available was that the cause of death was thought to be partial atelectasis. The main vessels branching off the arterial trunk were identified for us by the pathologist who performed the necropsy.

The congenital anomalies present in this heart have not previously been reported as occurring together. Defective development of the entire left side of the heart is present resulting in a hypoplastic left atrium, grossly stenotic left atrio-ventricular (A-V) opening, rudimentary left ventricle, and hypoplastic aorta. Associated with the left heart hypoplasia are a common brachiocephalic trunk, a bicuspid right A-V valve, and defective septum secundum development.

Anatomical Findings

The heart weighed 28 g., and the transverse diameter measured 5 cm. The external form appeared normal, except that no interventricular sulcus was present (Fig. 1). The visceral pericardium was normal. The heart had been cut open at the original necropsy, and at first sight it appeared to have two atria and a large single ventricle. The large ventricle proved, however, to be a right ventricle, and a rudimentary left ventricle was embedded in its wall.

Atria. The left atrium was much smaller than the right. Two pulmonary veins, one right and one left, entered the former, and the atrial wall had both a smooth portion and a portion lined with musculi pectinati. The musculi continued into a left auricular appendage, which almost encroached onto the anterior aspect of the heart.

In the floor of the left atrium there were two small depressions which appeared to end blindly. Under the dissecting microscope incisions were made vertically through these, into the ventricular wall. One of the depressions was found to lead through a grossly stenotic but not quite atretic canal (diameter 1 mm.) into a minute rudimentary left ventricle, about 2 mm. × 2 mm. × 2 mm., embedded in the muscular wall of the right ventricle. Valve cusps were absent from the A-V canal.

The left atrium communicated with the right atrium through two small defects in a septum primum. The larger defect (Fig. 2) was situated in the upper part of this septum and the smaller in the lower. On the right side of the septum primum a single band or cord extended across the right atrium antero-posteriorly, about 4 mm. to the right of the septum. The cord (Fig. 3) was attached to the posterior wall of the right atrium (on the left of the superior vena caval opening) and anteriorly to the left edge of the tricuspid ring. The cord was interpreted as being the rudiment of a defectively formed septum secundum, for reasons given below, and the openings in the septum primum as foramen primum and foramen secundum.

The right atrium received the superior and inferior venæ cavæ and, probably, the coronary sinus (*vide infra*), and communicated with the ventricle through an A-V valve. An auricular appendage with musculi pectinati extended anteriorly and to the left in an approximately normal position, while a sharp line of demarcation between the musculi pectinati and sinus venarum was present.

The coronary sinus, 10 mm. × 4 mm. in size, was identified in the coronary sulcus posteriorly, and communicated with an oblique vein of the left atrium. The coronary sinus was incised under the dissecting

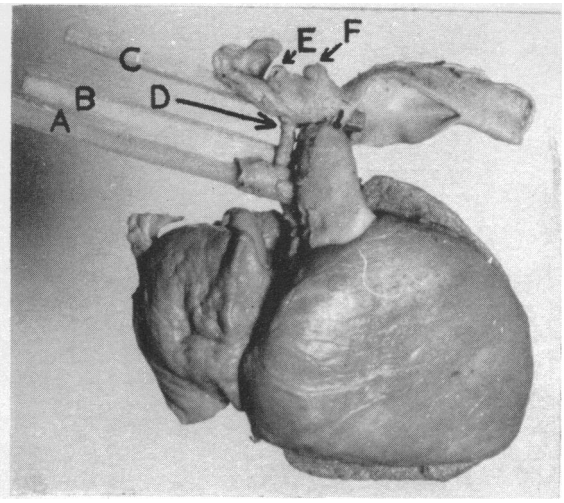

FIG. 1.—Anterior aspect of the heart. A and B=matchsticks in pulmonary arteries. C=matchstick in common brachiocephalic trunk. D=hypoplastic aorta. E=left common carotid artery. F=left subclavian artery.

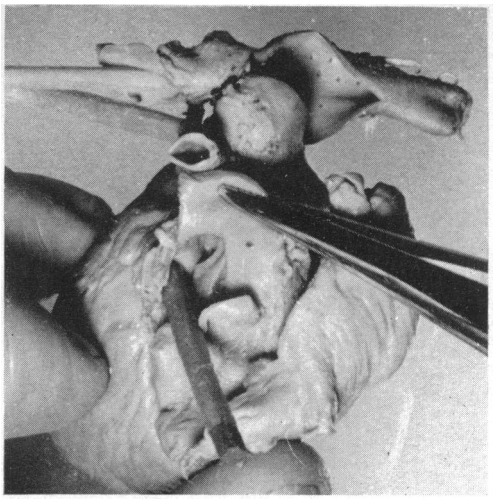

FIG. 2.—Left atrium. Matchstick through foramen primum.

microscope. Three small channels dipped deeply from the lumen towards the right atrial cavity. On probing with fine blunt needles, the actual openings into the atrium could not be found and were presumably of microscopic size. The site in the atrial wall where the probe could be felt through the endocardium was separated from the opening of the inferior vena cava by a small ridge, the sinus septum. This sinus septum projects in early embryos from the dorsal wall of the sinus venosus, intervening between the orifice of the vena hepatis communis and of the body of the sinus venosus. It may be identified intervening between the definitive inferior vena caval opening and the coronary sinus and its identity may be confirmed by the fact that it contributes to the formation of the medial end of the valve of the inferior vena cava (*Gray's Anatomy*, 1958). In this specimen the crista terminalis could be traced down into the lateral end of the valve of the inferior vena cava, and the medial end of the valve could be followed into the sinus septum.

In normal hearts, the anterior limb of limbus fossæ ovalis (the lower free border of septum secundum) bifurcates. The anterior division runs into the sinus septum as the superior limbic band, while the posterior one runs back to merge with the left venous valve (Hamilton, Boyd, and Mossman, 1952). In this heart, some fibres from the anterior end of the cord-like structure mentioned previously were traced, under the dissecting microscope, into the sinus septum, and others back along the atrial floor. The cord had the characteristic appearance of a defectively developed septum and this, together with the fact that it could be traced into the sinus septum, left little doubt that this band represented the malformed septum secundum.

Ventricles. The rudimentary left ventricle was lined with endocardium and contained blood clot in its lumen. It had no communication with the large right ventricle.

The greatly enlarged right ventricle was in communication with the right atrium through a large bicuspid right A-V valve, having only an anterior and a posterior cusp (Fig. 4). The right ventricular wall was 5 mm. thick at its maximum. A papillary muscle of the anterior cusp of the A-V valve was attached to the anterior wall. A papillary muscle of the posterior cusp was attached to the posterior wall, the attachment extending to the apex of the heart. Both cusps were attached at their superior angles by chordæ tendinæ to a thick trabecula carnea running horizontally across the posterior ventricular wall.

Arterial Trunk. The right ventricle communicated with a single outflow tract (Fig. 1), 8 mm. in diameter and guarded at its base by three well-formed valve cusps, posterior and right and left anterior. The heart had unfortunately been cut open through the valve ring at necropsy and the incision passed through a small flap of valvular tissue, which bridged between the left anterior and the posterior cusps. This might have been the remnant of a fourth, imperfectly formed cusp. The posterior cusp had minute perforations at both

FIG. 3.—Rudiment of defectively formed septum secundum (held in forceps).

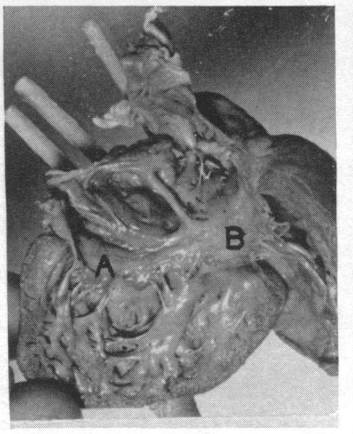

FIG. 4.—The two cusps, posterior (A) and anterior (B), of the right atrio-ventricular valve.

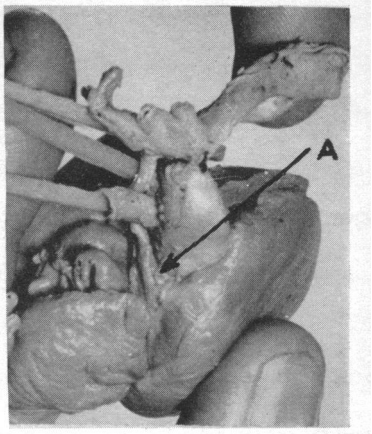

FIG. 5.—Hypoplastic aorta. A=division of the vessel. The anterior division can be seen running on to the anterior ventricular wall and giving off its branches.

its left and right ends. No coronary ostia were present at the base of this vessel, which is interpreted as a truncus pulmonalis.

The trunk ascended for 2 cm. and then arched to the left. The ascending part of the trunk gave rise from its right side to a vessel 4 mm. in diameter, with its ostium 6·5 mm. from the base of the trunk; and from its right side and slightly posteriorly, to a second vessel 3 mm. in diameter, its ostium being 7·5 mm. from the base. These two vessels were identified as the pulmonary arteries (Fig. 1).

The arch of the arterial trunk gave rise to a single vessel, an example of a common brachiocephalic trunk (Lev, 1953). This vessel passed superiorly and to the right and in turn gave rise to the following branches (Fig. 1 and 5).

(1) A slender vessel arose from the right and descended between the two pulmonary arteries. This closely resembled a coronary artery because of its calibre and the distribution of its branches. It is more correctly interpreted, however, as part of a grossly hypoplastic ascending aorta (Siddoway and Chernish, 1952). It ended by dividing at the level of the base of the trunk, on the anterior ventricular surface, into two divisions.

(a) The posterior division ran back for 2 mm. and divided into branches. One of these ran towards the rudimentary left ventricle, and presumably entered the ventricle: the continuity was disturbed, however, by the original incision into the heart, made at necropsy. The other branch ran backwards, downwards, and to the left over the posterior surface of the right ventricle, as a coronary artery ending at the apex of the heart.

(b) The anterior division ran forward for about 4 mm. and then divided into two branches. One of these turned to the right and entered the coronary sulcus, and the other ran to the left for a short distance over the anterior ventricular wall.

(2 and 3) From the left, two vessels arose, one at the level of the hypoplastic aorta described above and one 3 mm. further up. These two were identified as the left subclavian and common carotid arteries respectively.

(4 and 5) Finally the common brachiocephalic trunk divided into (4) the right common carotid and (5) the right subclavian arteries.

After giving off the common brachiocephalic trunk, the arch continued as the descending aorta which gave off intersegmental branches.

Discussion

The combination of left heart hypoplasia and common brachiocephalic trunk has been reported once previously (Oestreicher and Harris, 1950). The association of these anomalies with bicuspid

right A-V valve and defective development of the septum secundum in this case appears to be unique.

Where a single vessel arises from the ventricular portion of a heart, the diagnosis rests between true truncus arteriosus persistens and pseudotruncus (Collett and Edwards, 1949; Brown, 1950; Anderson, Obata, and Lillehei, 1957) of which there are two types. In the first, truncus aorticus, the aortic component of the original truncus arteriosus becomes dominant, while the pulmonary component regresses completely or remains as a small, hypoplastic vessel. In the second, truncus pulmonalis, the situation is reversed.

The hypoplastic ascending aorta of truncus pulmonalis is frequently mistaken for a coronary artery. The clue to its identity is obtained by tracing a small branch of the narrow vessel into the left ventricle (Siddoway and Chernish, 1952). Superiorly, the vessel arises from the aortic arch or one of its branches.

The diagnostic criteria that distinguish true truncus arteriosus from the two types of pseudotruncus have not in the past been clearly defined. Aortic hypoplasia, however, is frequently associated with hypoplasia of other left-sided structures in the heart, as in the example described in this report. Hypoplastic pulmonary trunk is similarly a frequent finding when right-sided structures are underdeveloped. In a separate communication (Blecher, 1962) the view will be presented that these associations may exist as the result of hæmodynamic factors in cardiac development. This suggests an additional criterion for diagnosing the three types of single arterial trunk, namely the presence or absence of hypoplasia in other parts of the heart. In the same paper a new classification of these and related disorders will be suggested, based on the same concept.

Summary

A heart is described in which a unique combination of congenital malformations was found at necropsy. Hypoplasia of left-sided structures (atrium, A-V canal, ventricle, and aorta) was present, associated with a common brachiocephalic trunk, bicuspid right A-V valve, and defective development of the interatrial septum.

The suggestion is made that hypoplastic development of different structures on the same side of the heart might be explainable in terms of hæmodynamic factors in cardiac development. Diagnosis might be aided by bearing this consideration in mind. This concept, and a classification based on it, will be more fully discussed elsewhere.

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References

Anderson, R. C., Obata, W., and Lillehei, C. W. (1957). *Circulation*, **16**, 586.
Blecher, S. R. (1962). To be published.
Brown, J. W. (1950). *Congenital Heart Disease*, 2nd ed., pp. 239–240. Staples Press, London.
Collett, R. W., and Edwards, J. E. (1949). *Surg. Clin. N. Amer.*, **29**, 1245.
Hamilton, W. J., Boyd, J. D., and Mossman, H. W. (1952). *Human Embryology*, 2nd ed. Heffer, Cambridge.
Gray's Anatomy (1958). 32nd. ed. Ed. T. B. Johnston, D. V. Davies, and F. Davies. Longmans, Green, London.
Lev, M. (1953). *Autopsy Diagnosis of Congenitally Malformed Hearts*, p. 64. Thomas, Springfield, Illinois.
Oestreicher, D. L., and Harris, G. E. (1950). *Amer. Heart J.*, **39**, 626.
Siddoway, J. L., and Chernish, S. M. (1952). *A.M.A. Amer. J. Dis. Child.*, **84**, 706.